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Interest of diffusion-weighted magnetic resonance imaging in IgG4 related renal disease detection and follow-up

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Introduction: Tubulointerstitial Nephritis (TIN) is a manifestation of IgG4-related diseases, which are characterized by infiltration of target organs by IgG4+ plasma cells and severe fibrosis. Cortico-sensitivity is one of the diagnostic criteria, but the treatment of steroid resistant and dependent forms is not well defined.

Case Report: We present a case of a 47-years-old patient with IgG4-related NTI followed for 72 months. He complained of fatigue and recurrent postprandial abdominal pain. With the exception of elevated levels of Gamma-Glutamyl Transferase (GGT), transaminases and IgG4, kidney function remained normal (serum creatinine ≤ 0.9 mg/dL). After 2 cures of methyl prednisolone (2010-11) azathioprine was associated in 2012. Due to the corticodependence and persistence of bilateral focal renal lesions detected by diffusion-weighted magnetic resonance imaging (DW-MRI), Rituximab (RTX) was given (2 × 376 mg/m²/15 days) in 2013. Before the first injection, Positron Emission Tomography (PET) showed metabolic hyperactivity corresponding to axillary and abdominal aorta lymph nodes but not in the kidney. After 4 months of RTX, the patient became asymptomatic. All biological alterations disappeared. PET showed a decrease in metabolic activity at extrarenal lesions described above. A dramatic regression of bilateral renal lesions was noted by DW-MRI: the apparent diffusion coefficient had almost doubled (0.776 vs. 1.111×10^{-3} mm²/sec) and the volume of renal lesions was reduced by 50%, which was never observed under other treatments.

Results: Our observations demonstrate: (1) the clinical, biological and radiological efficacy of rituximab in a steroid-dependent form of IgG4- related TIN and (2) the interest of DW-MRI as a non-nephrotoxic radiological and PET complementary approach not only in monitoring the effectiveness of immunosuppression but also in the early detection of renal involvement during IgG4 related disease.

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Successful completion of pregnancy using apheresis and a balanced dose of coagulation factors in the presence of high thrombophilia and Lp(a) levels in a woman with two previous abortions

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Importance: The case shows clinical impact of thrombophilia and Lp(a) increases in relation to unwanted abortions and a possibility of intervention to allow a successful pregnancy for affected patients.

Observations: This is a single case of a 26-year-old woman with two apoplectic insults at young age with different coagulation disorders (type 1 von Willebrand disease, protein S deficiency and increased levels of lipoprotein a) and also strong positive family history. Due to high active thrombophilia status and with respect to very thin coronary arteries a weekly lipoprotein apheresis treatment (LA) was initiated. Before LA was initiated the patient was getting pregnant twice and once under a weekly LA, but lost fetus each time. Under an intensified to treatment intervals twice a week she got again pregnant again. Pregnancy developed regular, without any complications. Baby was delivered in 34th week of pregnancy by caesarean section, developed well and could left the incubator from the 14th day after delivery.

Conclusions & Relevance: Under an intensive treatment regimen it was possible to allow a successful pregnancy for high thrombophilia and Lp(a) increased patient. This case shows the importance of an interdisciplinary and individual patient-centered approach in pregnancy management for affected women.

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